

Case Report

The first calcified acoustic neurinoma identified in China: a case report and literature review

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Abstract: Here we reported the first case of left cerebellopontine angle acoustic neurinoma with calcification in our department. The patient was 65 year-old, suffering from progressive loss of hearing in the left ear for about 30 years and headache with unsteady gait for approximately 6 months. Head CT & MRI scan identified an intracranial lesion located on left cerebellopontine angle. Left suboccipital retrosigmoid approach was applied to perform the operation after patient consent. The tumor was completely resected without complication and the patient recovered well. Histological findings revealed Spindle-shaped tumor cells tightly compacted to form the Antoni A region, while loosely arranged to form the Antoni B region. Hyaline degeneration and calcification formation were observed across the majority of the tumor.

Keywords: Acoustic neurinoma, calcification, cerebellopontine angle

Introduction

Acoustic neurinoma, meningioma and cholesteatoma are the most common tumors of the cerebellopontine angle region, representing 8% to 10% of all brain tumors. Among tumors of the cerebellopontine angle region tumors, neurinomas (>80%) and meningiomas (~15%) are most commonly observed. While 20% to 30% of meningiomas display calcification, these deposits, while extremely rare in neurinoma, can cause difficulty in radiological and histological diagnosis [3, 4, 6]. Fewer than 10 cases of acoustic neurinoma with calcification have been reported worldwide, and, until now, no single case has been reported from China. Herein we describe the identification of calcified acoustic neurinoma in our department from clinical and radiological image characters, clinical surgery and neuropathological features. Moreover in discussing this case report and performing literature reviews we hope to improve our knowledge and understanding of calcified acoustic neurinomas, to ameliorate diagnosis and treatment.

Case report

A 65 year-old male patient suffered from progressive loss of hearing in the left ear for about

30 years and headache with unsteady gait for approximately 6 months. Ten years ago, a further decrease in the patient's hearing led to his presentation at a local hospital for head CT scan, which identified an intracranial lesion located on left cerebellopontine angle (**Figure 1**). At that time, the patient declined further examination and treatment. In the past 6 months, the patient had headache and unsteady gait again representing at a local hospital for head MRI scan, which found a brain tumor located on left cerebellopontine angle (**Figure 2**). The tumor was obviously enlarged compared to previous imaging. At this time, the patient consented to further treatment.

Neurological examination

The patient was alert, GCS 15, bilateral pupils were round and equal with diameters of 3 mm, light reflex was sensitive. Muscle strength and tension was normal. Left hearing was lost, and left facial palsy with House-Brackmann grade I. Dystaxia with left extremities. Romberg's sign was positive, and Babinski sign was negative.

Surgical treatment

Left suboccipital retrosigmoidal approach was applied to perform the operation. During the

Calcified acoustic neuroma

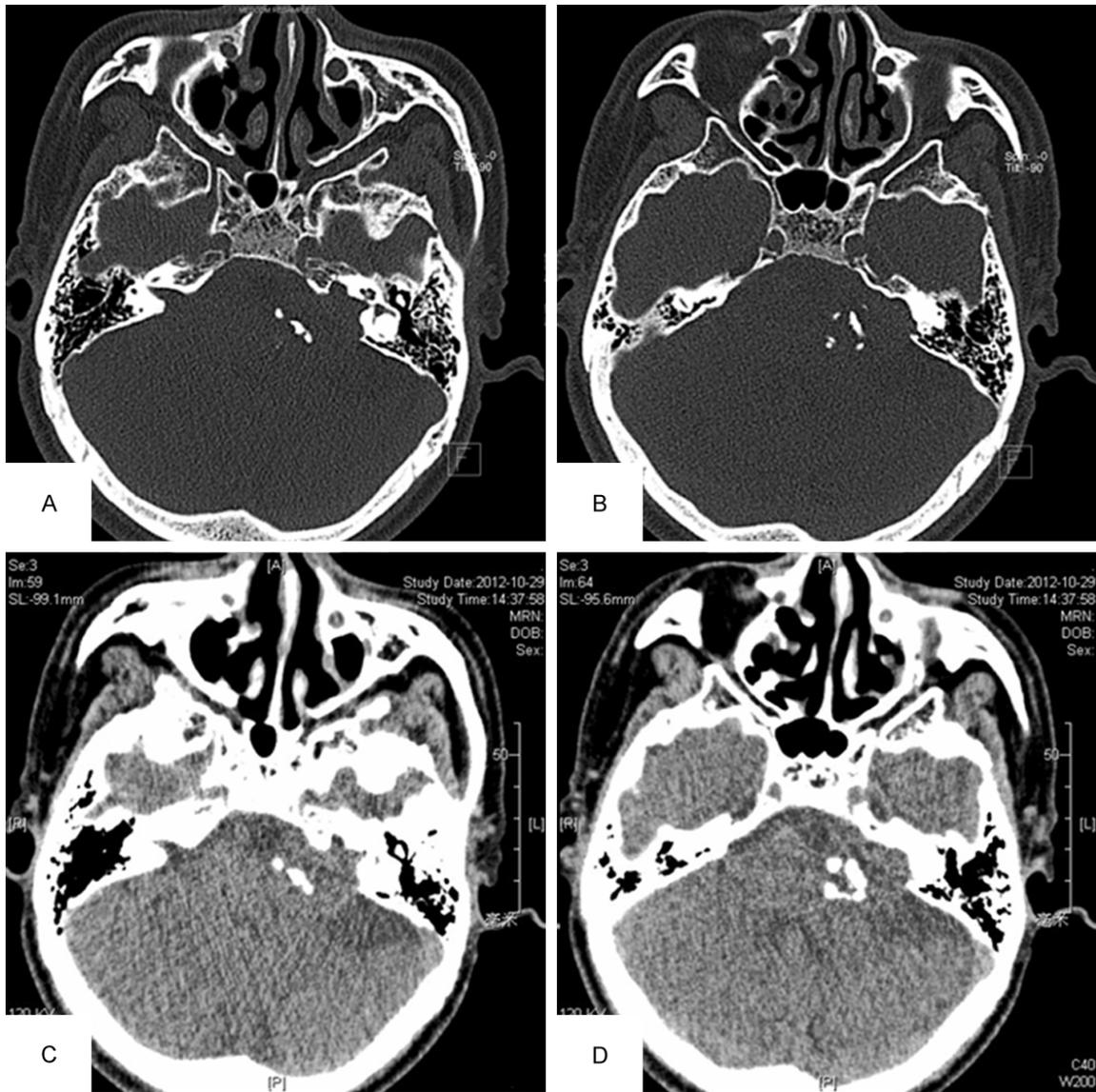


Figure 1. CT scan: a nearly round abnormal density lesion located in left CPA angle, about 3 cm×2.4 cm, boundary was clear with heterogeneous density. Several spots of high density calcification were found in the lesion. The brain stem and the fourth ventricle were compressed and the midline structure was shift to the right side. Left internal acoustic meatus was enlarged.

operation, the dura was opened and CSF was released to decrease the intracranial pressure. Cerebellum was elevated and the tumor was exposed. The tumor was grey white, stiff and tenacious (**Figure 3A**). The boundary was clear and the blood supply was abundant. After cutting open the tumor, calcification spots were observed in the tumor which was full of microvessels and microbleed structures (**Figure 3B**). The tumor was removed by pieces while the arachnoidal border was divided. As the intraoperative neurophysiological monitoring,

the facial nerve was detected at the ventral side of the upper pole of the tumor with the detect current of 0.35 mA, which was compressed thin by the tumor (**Figure 3C**). After removing the CPA part of the tumor, the internal acoustic meatus was drilled open to remove the small part of the tumor in it. Finally, microscopic total resection was achieved and the facial nerve was separated carefully and protected well. And then, the minimal response current of 0.05 mA was applied to verify the facial nerve's good function. The surgical pro-

Calcified acoustic neurinoma

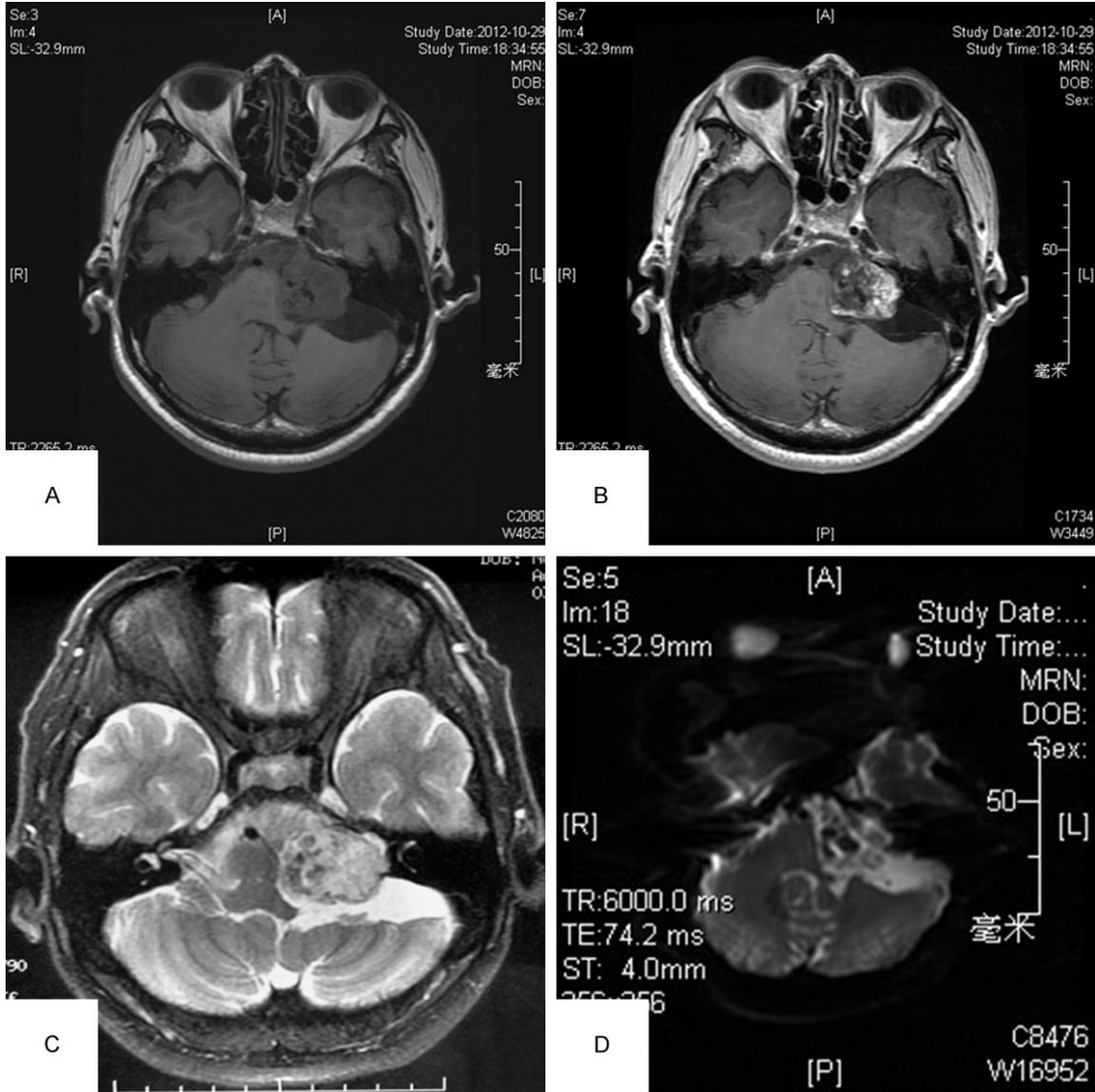


Figure 2. MRI scan: a nearly round abnormal signal lesion located in left CP angle, slightly long T1-weighted and long T2-weighted with spots of low signal lesion. The lesion was about 3.3 cm×2.4 cm with heterogeneous enhancement.

cedure was performed without complication and the patient has recovered well.

Pathological findings

Histological findings revealed Spindle-shaped tumor cells tightly compacted to form the Antoni A region, while loosely arranged to form the Antoni B region (Figure 4). Hyaline degeneration and calcification formation were observed across the majority of the tumor. Mild heteromorphosis was noted, while mitotic figures were not seen. There was no necrosis. Reticular fibers were pericellular (Figure 5). Immuno-

histochemical staining (Figure 6), the tumor cells were strongly positive for S100, Vimentin, CD56, and weakly positive for CD34. None of the tumor cells were immunoreactive for SMA, VEGF and EGFR, while the vascular endothelial cells were positive for CD34, SMA and VEGF. The MIB-1 index was about 1%.

Discussion

Calcified acoustic neurinomas are extremely rare with fewer than ten cases reported worldwide, and no single case report from a Chinese population until now. We described the rare

Calcified acoustic neurinoma

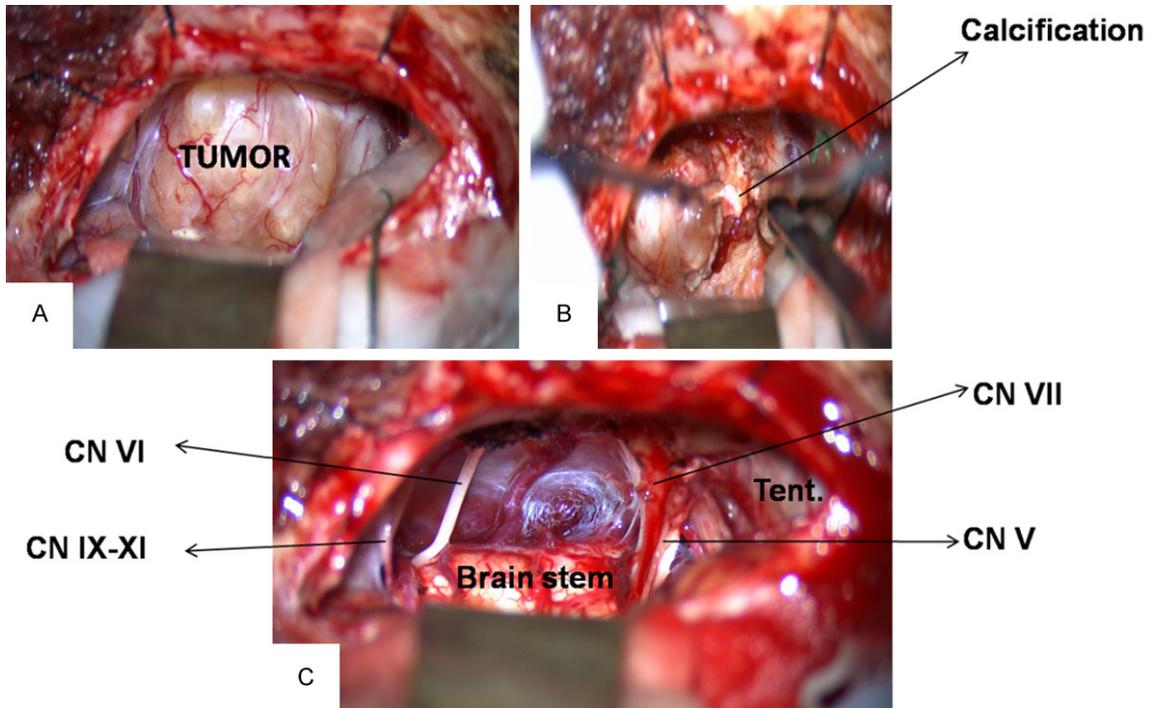


Figure 3. Intraoperative image. After elevation of the cerebellum, the tumor was exposed. While resecting the tumor, intratumoral calcification was observed. After total resection, the facial nerve, trigeminal nerve, abducent nerve and posterior group cranial nerve were all preserved well. The surface of the brain stem was smooth.

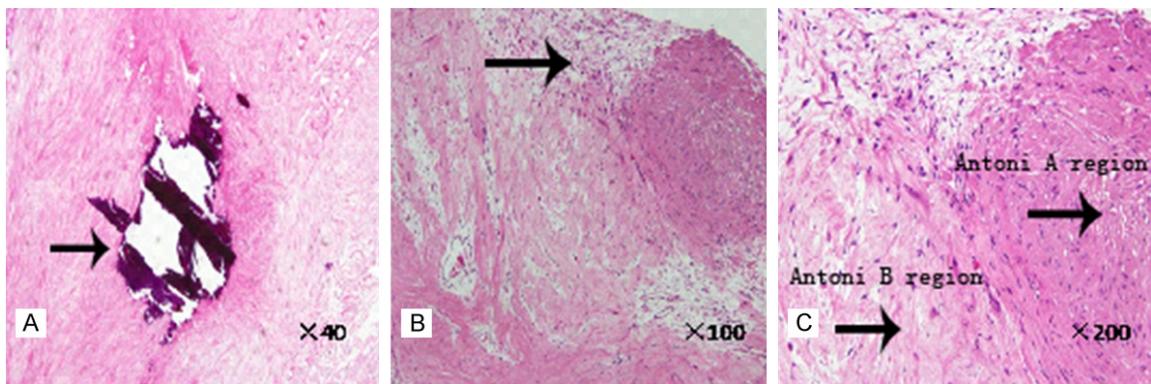


Figure 4. Photomicrographs of HE staining. A. Calcification and collagen sclerosis were observed across the majority of the tumor (as the arrow indicated; Original magnification $\times 40$). B. The tumor cells were seen in a small portion of tumor (as the arrow indicated; Original magnification $\times 100$). C. A medium-power image shows tightly compacted spindle-shaped tumor forming the Antoni A region, while loosely arranged to form the Antoni B region (as the arrow indicated; Original magnification $\times 200$).

identification of a patient who presented with a calcified acoustic neurinoma to raise more attention to the diagnosis of the calcified tumor in CPA region and the mechanism of the calcification. Clinical characters and pathological features of the calcified acoustic neurinoma were summarized. During immunohistochemical (IHC) workup, in addition to staining for: S100,

Vim, CD56, CD34, we also evaluated markers for microvascular genesis and reticular fibers staining.

In 1989, Beskin et al [1] reported a case of calcified neurinoma from the aspect of radiological image. He thought that 25%-30% meningiomas of CPA could have calcifications, and sug-

Calcified acoustic neurinoma

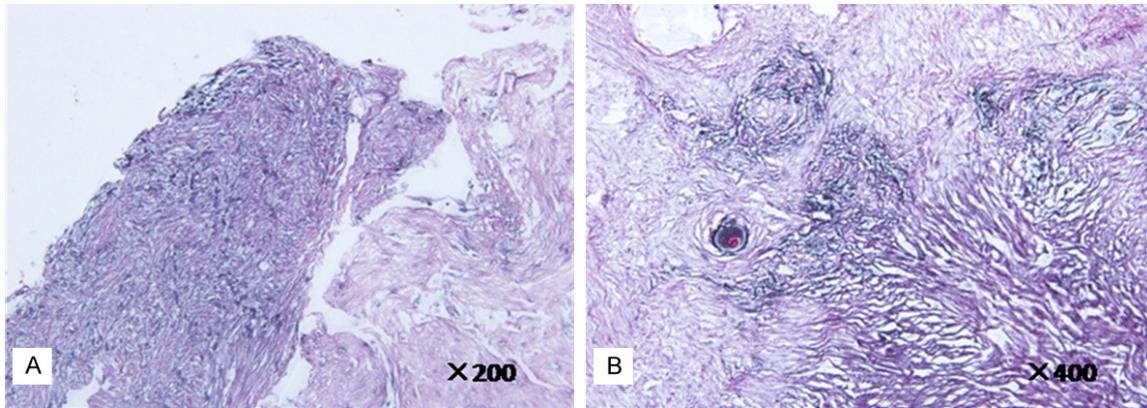


Figure 5. Reticular fiber staining showed that collagen sclerotic tissue was widely distributed in most of the tumor. (A, B) The tumor tissue was full of reticular fibers which displayed in black region.

gested that calcified tumors, especially with the enlargement of internal acoustic meatus, should be considered as acoustic neurinoma. In the present case, the calcified CPA tumor with typical enlargement of internal acoustic meatus, should be diagnosed as acoustic neurinoma. Thus, it is not difficult to make the exact diagnosis for the calcified tumor of CPA with enlargement of internal acoustic meatus.

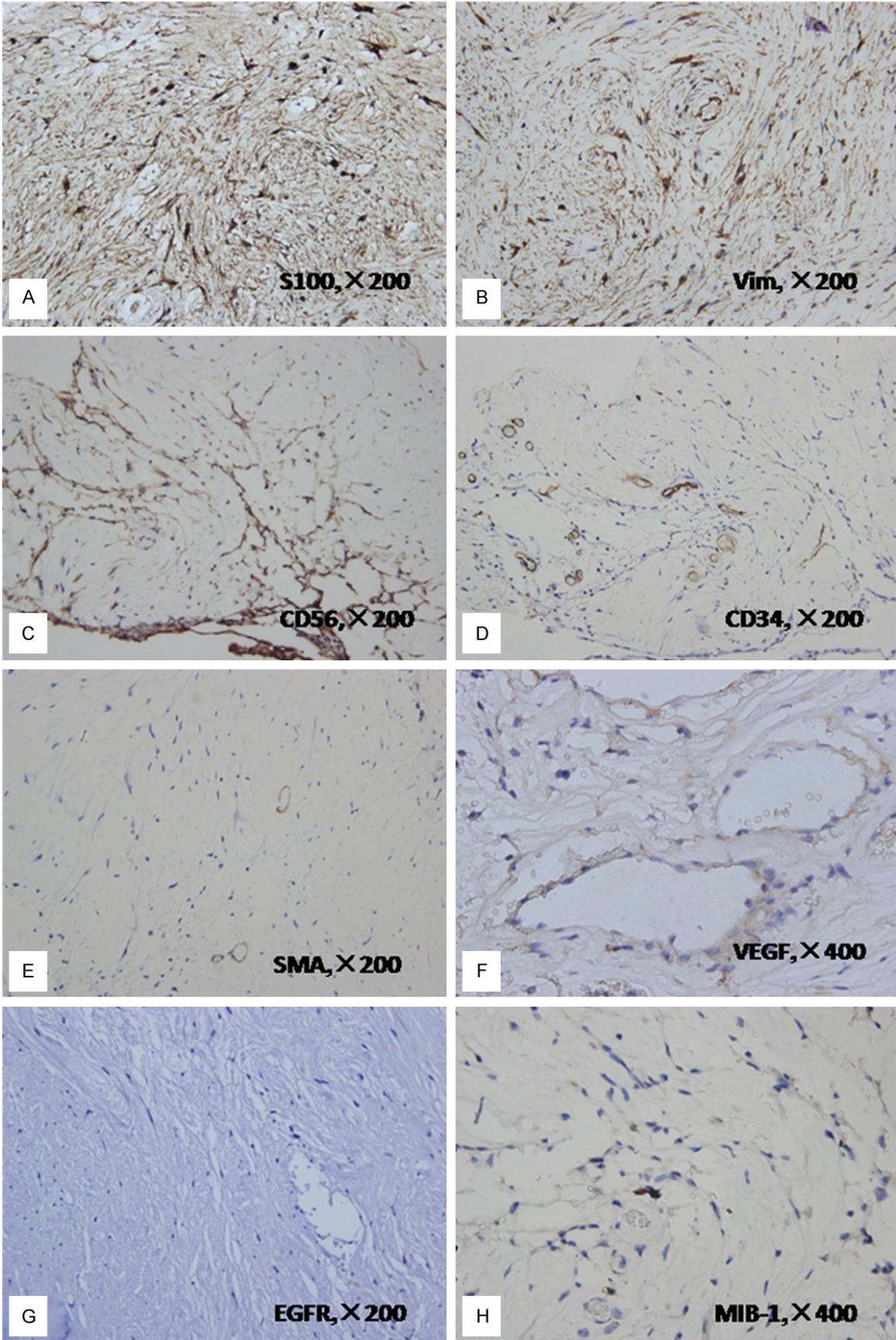
In 2002, Japanese scholar M. Tosaka [5] reported a case of calcified acoustic neurinoma with CD34 positive expression. They suggested spindle-shaped tumor cells gathered to form the cellular island, so called Antoni A region, which was the important histological character of Schwann neurinoma. The mechanism of calcification was not certain, but calcification of the acoustic neurinoma in their case might have resulted from ischemic or necrotic tissue related to minor and localized bleeding form abnormal intrinsic blood vessels. They also found that the spindle-shaped tumor cells of the sclerotic region (Antoni B region) were positive for CD34 immunohistochemical staining. In our case, we have found the similar phenomenon that the spindle-shaped tumor cells of Antoni B region had strong immunoreactivity for CD34. CD34-positive spindle-like cells may be related to the degenerative and fibrotic change of the tumor.

In 2011, Gopalakrishnan et al [2] reported two cases of acoustic neurinomas with calcification. Vestibular neurinoma was originated from the transmigration region between Schwann cells and oligodendrocytes of the vestibular

nerve. It was extremely rare for the calcification of acoustic neurinoma. Histopathological examination found that GFAP staining was negative while S100 staining was strong positive. They believed that the calcification of acoustic neurinoma was related to the rapid progression of the tumor resulted in ischemic necrosis and microhemorrhage. In our case, immunohistochemical analysis demonstrated that the staining for S100, Vim and CD56 were all strong positive in the spindle-shaped cells, which corresponded to the character of neurinoma. At the same time, we found immature microvessels structures in the tumor, which inferred that calcification might have relationship with ischemic necrosis resulted from microhemorrhage in the tumor. Reticular fiber staining demonstrated most part of the unstructured collagen region existed in the tumor, which inferred that collagen progression might be connected with calcification formation.

At present, the mechanism of calcification in acoustic neurinoma remains unclear, relating to: 1) the tumor history was very long (>10 years) suggesting calcification formation was a long process. 2) The tumor size was large, with a diameter ≥ 3 cm. It was inferred that the vascular supply could not correspond to the tumor growth, resulted in ischemic necrosis, and thus formed the calcification. 3) Abnormal microvessels structures in the tumor, resulted in microhemorrhage, and hematoma organization may lead to calcification. 4) A large quantity of unstructured collagen in the tumor, which suggested that the calcification might be the final result of the collagen progression.

Calcified acoustic neurinoma



Calcified acoustic neurinoma

Figure 6. Photomicrographs of immunohistochemical staining. A-C. The tumor cells are strongly positive for S100, Vimentin and CD56. D. The tumor cells are mild positive for CD34, while the vascular endothelial cells are strongly positive. E-G. The tumor cells were negative for SMA, VEGF, and EGFR. The vascular endothelial cells were positive for SMA and VEGF. H. The MIB-1 index was low, about 1%.

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Disclosure of conflict of interest

None.

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